

NOT JUST A STARTLE RESPONSE: ANESTHETIC MANAGEMENT OF A PARTURIENT WITH HYPEREKPLEXIA

Lindsey Nguyen, MD, Ashley Lewis, MD, Annastacia Woytash, DO, Holly Ende, MD, Taylor Ziga, MD

Department of Anesthesiology, Vanderbilt University Medical Center, Nashville, TN

CASE

- 31F G1P0 with hypererekplexia resulting in frequent falls, wheelchair use, and long-term anticoagulation
- Received regular prenatal care

- High risk obstetric anesthesia consult at 36w0d for PPRM
- High risk of hypertonic crisis
- Discontinued LTAC
- Continued home benzodiazepine
- Early DPE for labor analgesia

- > 24 hours of labor due to arrest of descent
- Underwent cesarean section under neuraxial anesthesia
- Pre-operative anxiolytic
- Nurse assigned noise control role

Postnatal course remained uncomplicated

BACKGROUND

- Hyperekplexia is a rare condition with incidence of less than 1:100,000
- Excessive startle response to auditory and/or tactile stimuli and electrolyte abnormalities
- Can lead to sudden death due to apnea, aspiration pneumonia, and cardiac irregularities

PATHOPHYSIOLOGY

- Involves mutation of the glycine receptor and presynaptic sodium and chloride-dependent glycine transporter genes
- Diminished muscle relaxation

TREATMENT

Anti-anxiety and anti-spasmodic drugs:

- Clonazepam
- Diazepam
- Phenobarbital
- Levetiracetam
- Phenytoin
- Sodium Valproate

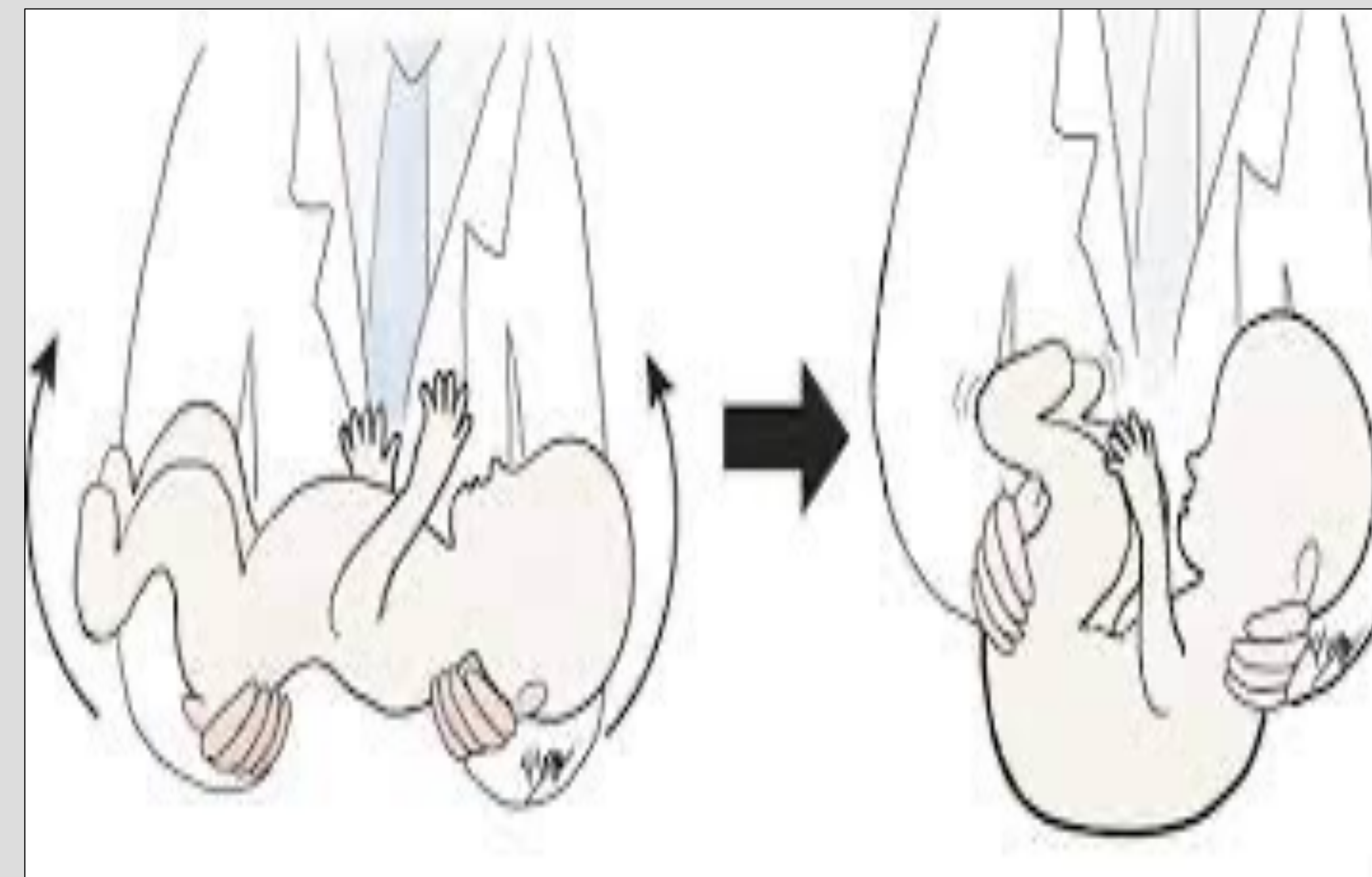
ANESTHETIC MANAGEMENT

PREVENTATIVE MEASURES

- Minimization of any unexpected stimuli and emotional stress
- Preemptive placement of neuraxial anesthesia to blunt intensity of contractions
- Anxiolytics

RESCUE MEASURES

- Vigevano maneuver



- Benzodiazepine administration

ANESTHETIC MODIFICATIONS

- Proactive treatment of hypotension
- Electrolyte repletion
- Lateral positioning for neuraxial placement
- Caution with succinylcholine and sevoflurane usage for general anesthesia

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